Trans-Cranial Placement of an Amplatzer Device to Control Intractable Cardiac Failure in an Infant with a Vein of Galen Anomaly

A Case Report

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Summary

Neonates presenting with intractable cardiac failure due to vein of Galen aneurysmal malformations (VGAMs) rapidly progress to multisystem organ failure and death if left untreated. Currently the only viable treatment option is endovascular embolization. Although intracranial embolization of a neonate is a high-risk procedure, successful treatment can reverse cardiac failure and prevent neurological complications associated with VGAMs. Embolization via the arterial route is thought to have a better outcome than embolization via the venous system. However, multiple transarterial embolizations in different sessions may well be contraindicated in neonates, because repeat access via the femoral artery, carries a risk of arterial trauma which, in turn, can jeopardize lower limbs. With this case study we show that after repeat failure of arterial embolization, the transcranial placement of an Amplatzer PFO occluder (AGA Medical, Plymouth, USA) in the aneurysm can effectively reduce intrafistular pressure and venous outflow velocity. We also propose a mathematical model that can be used to calculate flow velocity through the aneurysm, which, in turn, could be used to aid clinical decision-making. Unlike some conventional techniques, the placement of an Amplatzer occluder does not pose the risk of completely obstructing venous drainage and therefore does not increase the risk of venous breakthrough hemorrhage. We propose this endovascular technique as a treatment option for high risk neonates in need of emergency embolization of VGAMs, where multiple arterial embolizations failed to control the condition sufficiently.

Introduction

A vein of Galen aneurysmal malformation (VGAM) is a congenital anomaly of the cerebral vasculature that alters the arrangement of blood flow in the deep venous system of the brain and significantly affects cerebrospinal fluid flow and production [1-4]. The anterior and posterior choroidal arteries as well as the anterior cerebral artery normally drain via mesencephalic collateral veins, but in the presence of a VGAM arterial blood drains directly into the median prosencephalic vein of Markowski. The procencephalic vein of Markowski is the embryonic precursor of the vein of Galen that, in rare cases, persists to form an enlarged venous

pouch [1,3]. The size of the venous pouch correlates with the severity of clinical symptoms [3,5,6]. Large aneurysms sequester a considerable amount of arterial blood. The "stealing" of arterial blood detriments the surrounding brain parenchyma causing "melting brain syndrome" [2,5], and necessitates a compensatory increase in blood volume which causes pulmonary hypertension. Pulmonary hypertension coupled with the increased venous return and the associated enlargement of the right atrium result in high-output right ventricular cardiac failure [2,5,7,8].

Neonates with VGAMs presenting with high-output cardiac failure have negligible chances of survival without endovascular embolization [2,9,10]. Here we report a case of an infant, with a choroidal type VGAM that presented with massive intractable cardiac failure, which we successfully treated with the transcranial placement of an Amplatzer occluder.

Case Report

Clinical information

A 3.92 kg boy, born by elective caesarean for breech position, was healthy at birth (Apgar score 8, 9 and 10 at one, five and ten minutes post delivery, respectively) but developed acrocyanosis 12 hours later. Cardiac ultrasound showed increased pulmonary pressures and severe tricuspid regurgitation. A bruit in his head prompted CT angiography of the brain. This showed a vein of Galen aneurysmal malformation (VGAM), but no visible brain damage. On his second day of life the baby developed congestive cardiac failure. Despite aggressive pharmacokinetic treatment his condition worsened. He was referred to our neurointerventional unit for emergency endovascular embolization at seven days of age. (The author PAF has successfully embolized 15 infants with VGAMs over the past ten years.)

Intervention

A cerebral angiogram done by femoral artery approach demonstrated multiple enlarged choroidal arteries as well as the posterior part of the pericallosal artery to supply the 25 mm diameter vein of Galen aneurysm. Blood from the VGAM drained into the straight sinus with high blood flow (Figure 1A). Eight of the multiple vessels, supplying the aneurysm, could be embolized transarterially during two separate procedures. For the first procedure we approached embolization from the left femoral artery. Catheterization of the left vertebral artery as well as the right carotid and left carotid arteries were done with a 4 French Headhunter catheter (Glidecath, Terumo, Japan), and the choroidal arteries, supplying the aneurysm, were superselectively catheterized with a micro-catheter (Echelon, Micro Therapeutics, Irvine, USA). Embolization of four arterial feeders was done with 90% histoacryl (N-Butyl Ester of A-Cyanoacrylate) with tantalum powder and a drop of lipiodol (Guerbet, Aulnay-Sous-Bois, France). Sixteen days later, we approached embolization from the right femoral artery. We again accessed the site of the aneurysm with a 4 French Headhunter catheter (Glidecath, Terumo, Japan) and microcatheter (Tracker Excel, Boston Scientific, Massachusetts, USA) and embolized another four arterial feeders using 80% histoacryl (N-Butyl Ester of A-Cyanoacrylate) with tantalum powder. For each procedure 22 cc contrast-medium (Jopamiron 300, Schering, Berlin, Germany) were injected over a period of 3 h - the maximum volume tolerable as indicated by body mass. Consequently, each procedure was limited to the amount of contrast medium that we could administer without risking the development of acute pulmonary edema.

Although the embolizations were successful, it did not reverse cardiac failure. At three months of age the baby was still ventilated and in a critical condition. Because transarterial embolization seemed ineffective, and because arterial access posed the risk of arterial trauma, which would jeopardize the lower limbs, we decided to transvenously embolize the VGAM. At the outset of the transvenous procedure we gained access to the aneurysm by placing a 4 French sheath (Cook Medical, Bloomington, USA) in the femoral vein and a 4 French Cobra 2 (Terumo, Japan) as a guiding catheter into the straight sinus. We attempted to deploy micro coils in the VGAM with the use of a micro catheter (Rebar-14, Micro Therapeutics, Irvine, USA) inside the guiding catheter. However, the VGAM had a morphological widening on the posterior side and this, coupled with the severe high flow pressures through the aneurysm, prevented the coils from staying in position.

It was evident that we were not going to achieve a good outcome with embolizing agents normally used for VGAMs, and we opted to use an unconventional approach and an Am-

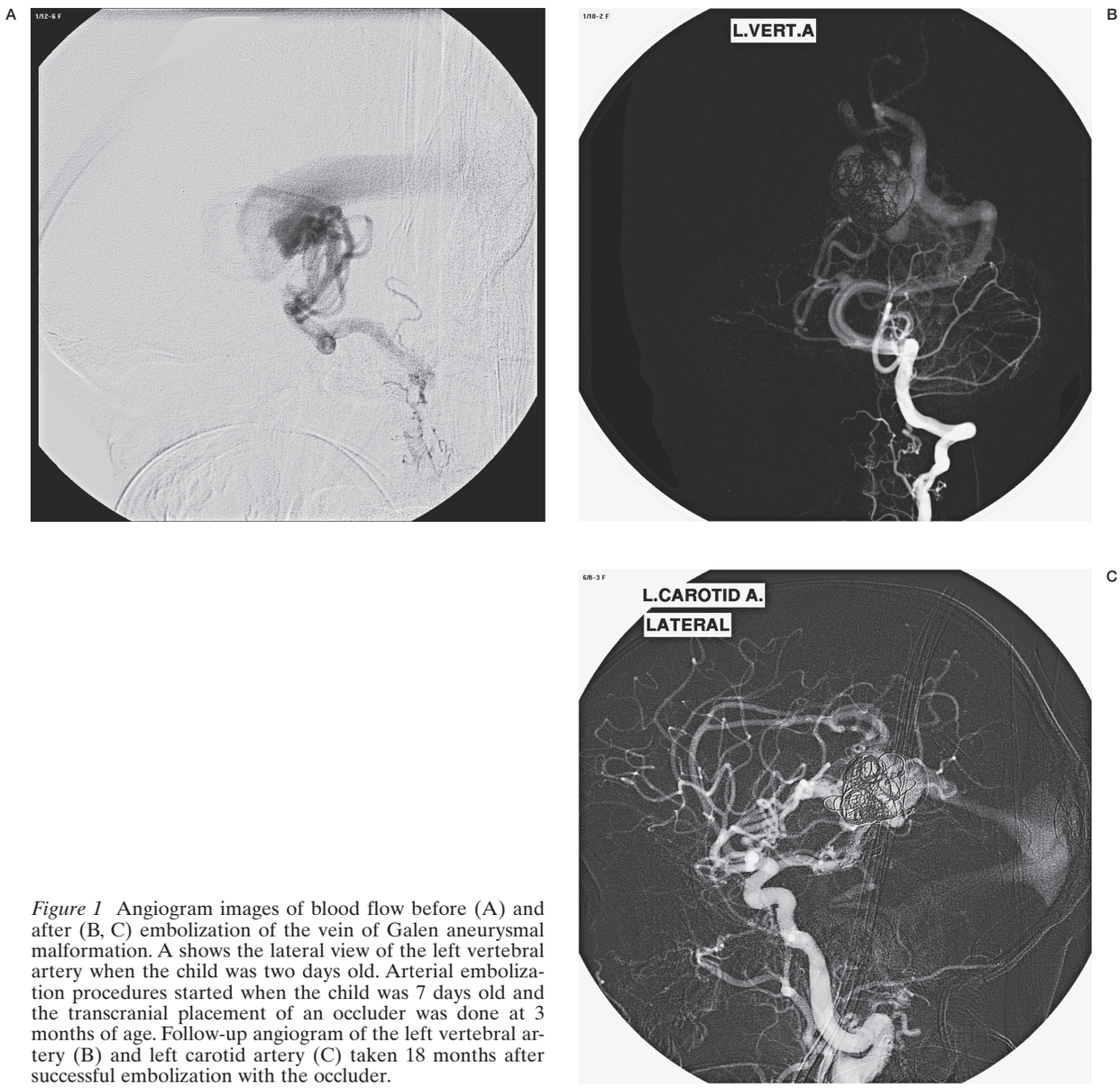
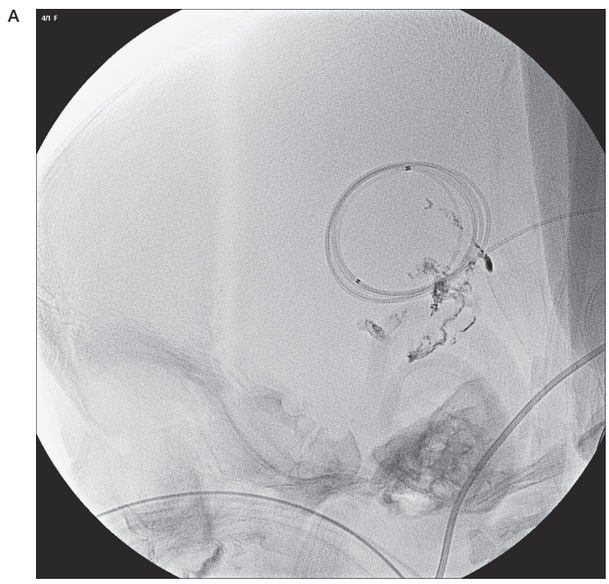


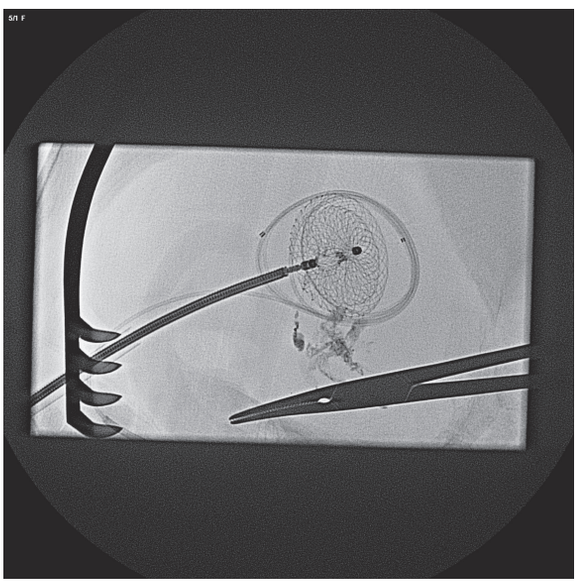
Figure 1 Angiogram images of blood flow before (A) and after (B, C) embolization of the vein of Galen aneurysmal malformation. A shows the lateral view of the left vertebral artery when the child was two days old. Arterial embolization procedures started when the child was 7 days old and the transcranial placement of an occluder was done at 3 months of age. Follow-up angiogram of the left vertebral artery (B) and left carotid artery (C) taken 18 months after successful embolization with the occluder.

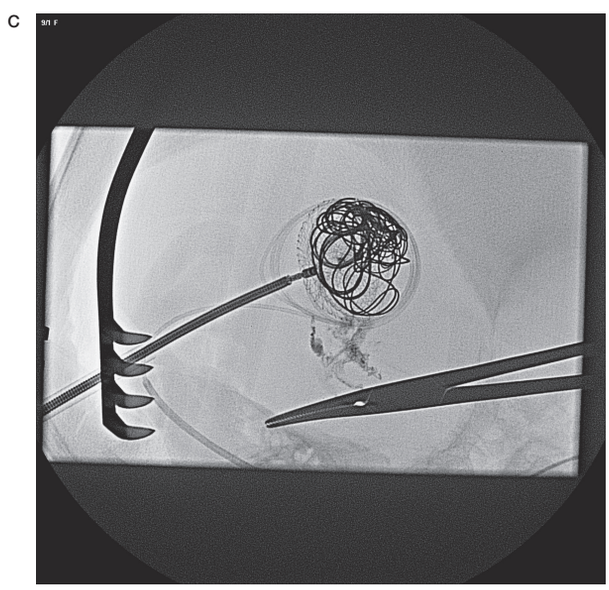
platzer PFO occluder (AGA Medical, Plymouth, USA). First the microcatheter, positioned in the aneurysm for our initial transvenous attempt, was coiled into the aneurysm to form a ring, as shown in Figure 2A. Thereafter the baby was discharged from theatre. The microcatheter offered enough resistance to blood flow through the aneurysm, and the baby's pulse rate lowered, from 188 to 120 b/min over the eight hours.

Placement of the Amplatzer was performed eight hours after the microcatheter was introduced into the aneurysm. A neurosurgeon gained access into the straight sinus via a small borehole just left of the torcula, where a micropuncture needle (Cook Medical, Bloomington, USA) was placed into the left transverse sinus. Through this a nitinol guidewire (Cook Medical, Bloomington, USA) was placed and then a dilator was introduced over the guidewire into the VGAM. This was followed by a 9 French sheath (Cordis Corporation, Bridgewater, USA). The occluder was introduced with an 8 French sheath pushed through the 9 French sheath.

The occluder is a self-expanding double-disk







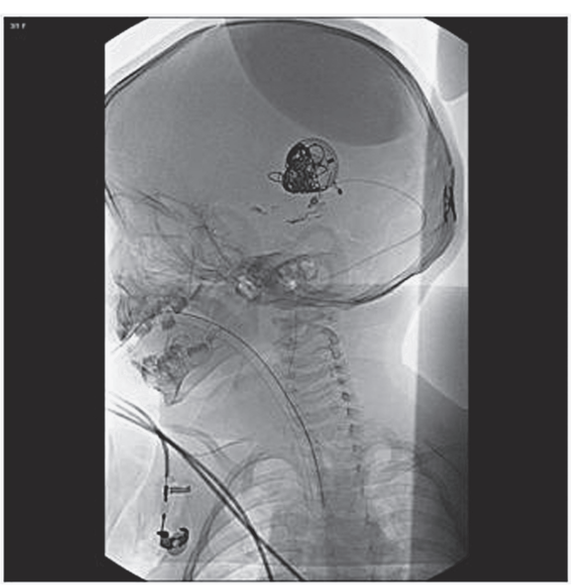


Figure 2 Embolization of the vein of Galen aneurysmal malformation with an Amplatzer PFO occluder. A) Coiling of the microcatheter into the aneurysm. B) Introduction of the occluder. The occluder consists of two wire mesh disks that are connected in the middle. The disks were positioned on either side of the microcatheter ring. C) The position of the microcoils.

Figure 3 The systemic part of the microcatheter was left in place for stability of the occluder and the coils for about 8 months. This angiogram was taken when the microcatheter was removed without mishap via a small jugular venotomy.

nitinol wire mesh with a connecting "waist" made from dense polyester fabric. Thus, the device obstructs central blood flow by means of its solid core, whilst allowing blood flow at the periphery. The occluder was deployed through the coils of the micro-catheter in such a way that the two disks of the occluder positioned on either side of the micro catheter ring (Figure 2B). This secured the occluder into the outflow tract of the aneurysm. Five microcoils (QC-16-30-helix and QC-8-30-helix both Micro Therapeutics, Irvine USA, GDC 18 (X2) and GDC 10, both Boston Scientific, Massachusetts, USA) were deployed via the microcatheter on

the upstream side of the occluder to further increase resistance to central outflow (Figure 2C). The angiogram done immediately post embolization showed a marked decrease of blood flow through the aneurysm.

The baby suffered from a slight fluid overload in the 12 hours following embolization, but this was successfully managed with Furosemide. Eight days post embolization the external part of the microcatheter was cut at the skin of the groin, and the rest left inside the vein. The systemic part of the microcatheter was left in place for stability of the occluder and the coils for about eight months, where after it was removed without mishap via a small jugular venotomy (Figure 3).

Mathematical model of blood flow dynamics before and after embolization

The change in blood flow dynamics between the choroidal arteries and the VGAM after embolization can possibly be explained by intravascular pressure adjustments.

To calculate the resistance that is needed to successfully reduce venous outflow, we propose the following equations which represent flow in a pipe system, including VGAMs.

Symbols used in the equations are defined as follows:

A = cross sectional flow area of the artery;
d = diameter;
K_R = resistance;
Q = volumetric flow rate;
u = linear velocity and;
ρ = density (blood has a known value of 1060 $kg.m^{-3}$)

The variation in arterial and venous pressure (ΔP) provides an energy driving force for blood flow, and the linear velocity of this flow is directly related to the volumetric flow rate according to *equation 1*:

$$Q = uA(m^3/s)$$

where the cross-sectional flow area of the artery is calculated according to *equation 2*:

$$A = \pi (d^2 \times 0.25)$$

Since the diameter of the supplying artery remains relatively constant, a reduction in total flow rate in the dilated vein and an increase in resistance to flow in the aneurysm can be explained according to *equation 3*:

$$\Delta P = K_R (\rho u^2 \times 0.5)$$

An increase in frictional resistance of blood flow through the VGAM will thus reduce flow velocity in the choroidal arteries, enhancing parenchymal perfusion.

Follow-up

Six months after therapeutic intervention MRI of the brain showed a reduction in ectatic vein diameter, a reduction in the number of venous channels and an obvious reduction in the size of the VGAM. No new abnormalities had developed. The ventricles showed moderate dilatation. Echocardiography showed dilated pulmonary arteries, slight ventricular dilation but normal left ventricular function and normal pulmonary venous return, a trivial tricuspid valve regurgitation and a small (>3mm) patent foramen ovale. There were no pericardial effusions, no vegetations, no signs of cardiac failure and normal coronary arteries. According to pediatric neurological assessment, the baby's cognitive development was normal, his hand-eye coordination symmetrical, and his hearing and eyesight normal. Figures 1 B and 1 C also show follow-up angiogram of left vertebral and left carotid artery done 18 months post embolization with the occluder.

Informed consent was obtained from the child's parents for all imaging studies, endovascular procedures and developmental assessments.

Discussion

Endovascular embolization is a viable treatment option for the long-term management of high-flow VGAMs [2,11-13]. Despite considerable advancements in the endovascular management of children with this abnormality, morbidity and mortality remains high among neonates with VGAMs that present with high-output cardiac failure [13-15].

Currently the optimal age for embolization of high-flow VGAMs is thought to be four to six months of age because the risk of the endovascular procedure is weighed against the development of cerebral damage [7,16,17]. Initial treatment is directed at reducing blood volume with diuretics and increasing cardiac contractility with inotropic agents in an effort to moderate cardiac insufficiency ahead of endovascular embolization [18,19]. However, in some neonates

pharmacokinetic control of cardiac failure is not possible and urgent endovascular embolization is required [13,14]. Other clinical symptoms that negate the delay of embolization are the early development of hydrocephalus, a rapid increase in head circumference, the development of encephalomalacia, evidence of arterial steal, and progressive occlusion of the venous outflow [7,16].

Embolization of the VGAM via the arterial route is thought to have the best outcome [5,19]. However, in neonates with intractable cardiac failure, arterial embolization is particularly difficult because limited amounts of parenteral fluid, such as contrast medium, can safely be administered and multiple arterial punctures may cause femoral arterial trauma. This coupled with the extreme tortuosity of the neonatal arterial system make arterial embolization a very high-risk procedure [5,20]. In some cases, the venous route poses easier access to the aneurysm, but it is perceived to have a higher risk than transarterial procedures because complete occlusion of venous outflow can cause breakthrough intraventricular bleeds, cerebral venous infarctions and a seemingly higher percentage of neurological complications on follow-up assessment [7,15].

With this case study we show that in neonates, VGAMs can be embolized successfully with the transcranial placement of an Amplatzer PFO occluder within the venous pouch of the aneurysm. The use of these occluders is well known in pulmonary arteriovenous fistulas, but as far as we could establish this is the first ever use of such a device for a VGAM. The design of the Amplatzer PFO occluder provides resistance to central blood flow through the aneurysm, without the risk of completely occluding venous drainage via the straight sinus and veins entering the vein of Galen from basal ganglia.

The neurodevelopmental outcomes of these infants are generally considered to be poor. In addition, they often rapidly progress to multiorgan failure and consequently score so low on the Bicêtre scoring system* that they are sometimes deferred for treatment. In this case, the patient scored 8 on assessment 48 h after birth.

Our patient's good neurological outcome confirms that these neonates can have normal neurological development if embolization obliterates the aneurysm soon after birth. This is supported by Mitchell et al. [13] and Frawley et al. [14] who reported normal neurological outcomes in 66% of patients in whom embolization was performed before four months of age.

We propose this novel therapeutic approach as a treatment option for neonates with a VGAM in need of emergency endovascular embolization and even for those that develop congestive cardiac failure soon after birth. The risk associated with transvenous embolization of VGAMs in neonates with an occluder, may well be less than the risk associated with embolization using traditional agents. If this is the case, the current practice that denotes delay of endovascular treatment of neonates with high-flow VGAMs, could be re-evaluated.

An important motivation for the use of a safer embolization technique is the serious neurological sequealae of the high cerebral venous pressure associated with VGAMs. In infants high venous pressure leads to hypoxia and obstruction of venous outflow as well as a possible imbalance of cerebrospinal fluid causing hydrocephalus and cerebral edema. These complications cause cognitive impairment and mental retardation due to progressive cerebral parenchymal damage [7,16]. Bansal et al. (2009) suggest that neurological damage is irreversible and progressively regresses even after successful embolization. The medical management of these neonates, coupled with the waiting period impose the potential for irreversible neurological damage and this may possibly be the basis for the poor prognosis regularly reported for neonates with high flow aneurysms which were stabilized medically whilst awaiting embolization at four or five months of age. For this reason we think treatment of neonates with congestive cardiac failure should be viewed as a matter of urgency, regardless of age.

In conclusion, with this case study we propose a safer endovascular procedure which may well shift the balance of therapeutic risk in favour of earlier treatment in lieu of the development of cerebral damage. Although transarterial embolization is considered ideal, in neonates a transvenous approach is expedient, and we think it may possibly contribute to good neurological outcome.

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